Moyamoya Syndrome: Post Cranial Irradiation of Pineal Gland Tumor

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Key words: Moyamoya syndrome, pineal tumor, radiation therapy

Summary

A right-handed eight-year-old boy, with headache, vomiting and positive parinaud 's sign was diagnosed as having a pineal gland tumor which histopathological section from surgical biopsy revealed to be a germinoma. The patient underwent ventriculoperitoneal shunt for obstructive hydrocephalus. Thereafter, he received cranial irradiation as definitive treatment. He was well and went back to school until five years later he developed a transient ischemic attack. Cranial magnetic resonance imaging showed a complete cure of the pineal tumor without any other specific abnormality. Eight months later he had an episode of stroke which was demonstrated by cranial computed tomography as acute left cerebral infarction in the middle cerebral artery territory. Cerebral angiography showed Moyamoya syndrome.

Introduction

Moyamoya-like syndrome and moyamoya disease are rare cerebrovascular occlusive diseases characterized by the angiographic appearance of progressive large-vessle vasculopathy with collateralisation [1,2].

The abnormal vascular network at the base of the brain is known as moyamoya vessel [3,4,5,6]. Moyamoya syndrome differs from moyamoya disease as it has one or more of these angiographic patterns such as asymmetrical involvement, loss of transdural angiogenesis and abnormal vasculature of the posterior fossa.

There are several reports of moyamoya syndrome after radiation of brain tumors such as hypothalamic chiasmatic glioma [7], optic glioma [8], pituitary adenoma [9] and suprasellar germinoma [1].

In this study, we report a case of moyamoya syndrome occurring four years later in a patient who received cranial irradiation for germinoma in the pineal region.

Case report

A 12-year-old boy presented with sudden onset of right hemiplegia, facial palsy and aphasia that developed immediately after playing football with his friends.

History: he was diagnosed with a pineal tumor at age 8 years. Plain radiograph of the skull showed prominent calcification in the pineal region (figure 1). Cranial computed tomography showed a mixed density of the pineal mass with central nodular calcification (figure 2). Cranial magnetic resonance (MR) imaging showed a well circumscribed isointense T1-weighted (figure 3A), hyperintense T2-weighted mass with increased enhancement of the pineal tumor after intravenous gadolinium ad-

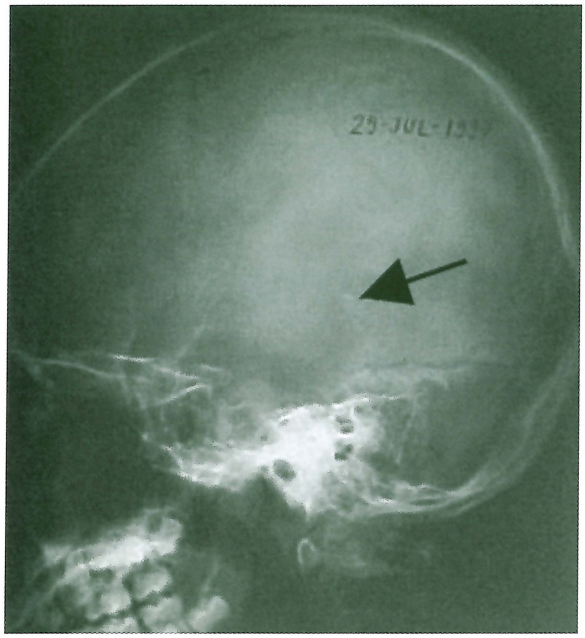

Figure 1 Lateral skull radiograph revealed a clump of intracranial calcification (arrow) in the pineal region.

ministration (figure 3B). Surgical tissue biopsy revealed germinoma. He underwent ventriculoperitoneal shunting for obstructive hydrocephalus. Radiation therapy of the brain was performed one month after surgical biopsy with a daily radiation dose of 180 rads for 31days (total dose = 5,620 rads) which comprised whole brain irradiation (3,420 rads) and additional localized pineal region radiation (2,200 rads). He was well and went back to school for five years. Thereafter, he frequently had transient right hemiparesis and facial palsy especially after exercise. Each attack lasted 2-3 h. The second cranial MRI showed no residual or tumor recurrence (figure 4A-D). Tumor markers (CEA, alpha-fetoprotein, beta-HCG) were in normal range. He was closely followed up monthly until eight months later when he had a stroke.

The CT examination disclosed a large area of low attenuation in the left fronto-temporo-parietal lobes, suggestive of acute cerebral infarction in the left middle cerebral artery territory. Physical examination of the cardiovascular system including chest radiograph, electrocardiogram and echocardiogram were normal. The patient underwent cerebral angiography (figure 5A-D) which demonstrated complete occlusion of the left internal carotid artery (ICA) at the supraclinoid portion of the carotid artery without filling of contrast medium in the left anterior cerebral artery (ACA) and middle cerebral artery (MCA) (figure 5A). Narrowing of the supraclinoid part of the right ICA and

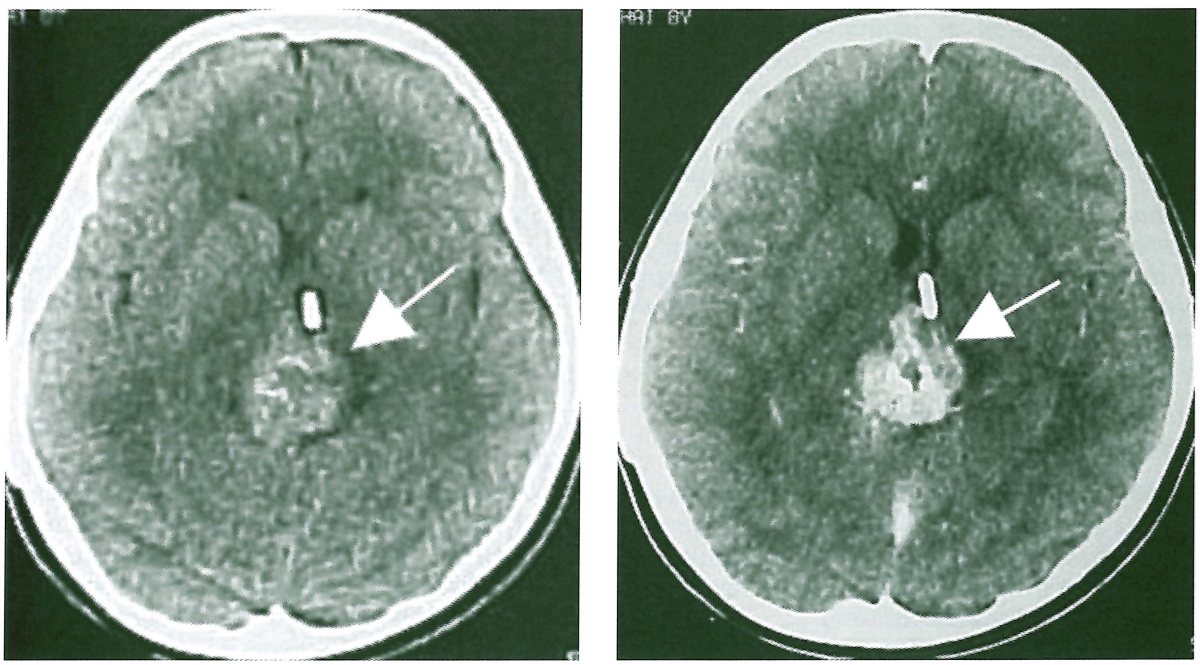

Figure 2 Axial pre and post contrast enhanced CT scan showed a well circumscribed enhancing mass (arrow) in the pineal region, germinoma. Tip of the ventriculoperitoneal shunt was noted in the left foramen of Monro.

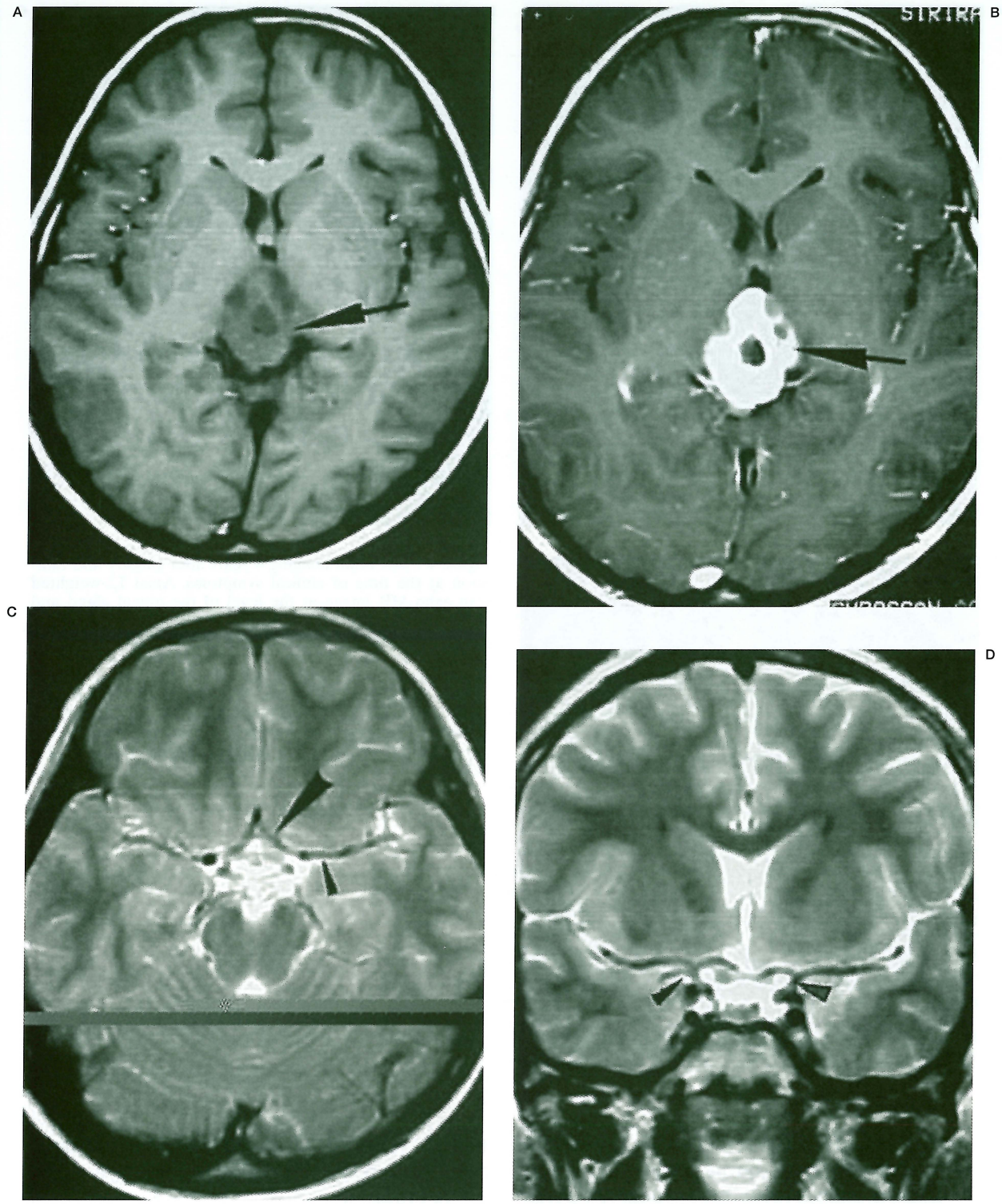

Figure 3 MR brain imaging prior to treatment. Axial T1-weighted (A) and T1-weighted after intravenous gadolinium administration (B) spin echo MR images showed an isointense mass (arrow) in (A) with central hypointense calcification and homogenous enhancement after gadolinium contrast study. Axial T2-weighted spin echo MR image (C) at circle of Willis showed normal signal void of normal patency of A1 segment (large arrowhead) and M1 segment (small arrowhead). Coronal T2-weighted spin echo MR image (D) also demonstrated the normal supraclinoid portion of bilateral ICA (arrowheads), normal signal of the basal ganglia and thalamus.

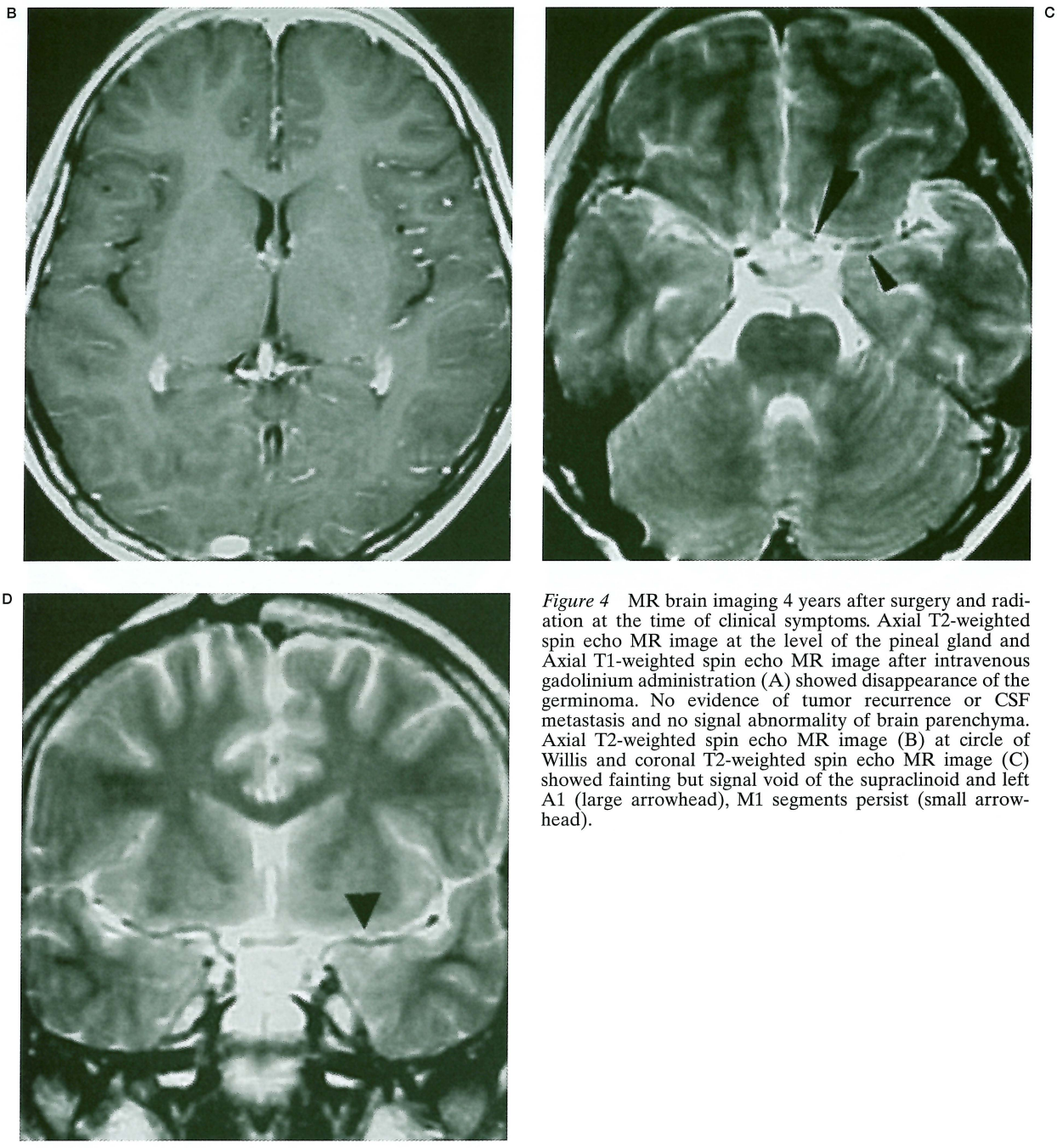

Figure 4 MR brain imaging 4 years after surgery and radiation at the time of clinical symptoms. Axial T2-weighted spin echo MR image at the level of the pineal gland and Axial T1-weighted spin echo MR image after intravenous gadolinium administration (A) showed disappearance of the germinoma. No evidence of tumor recurrence or CSF metastasis and no signal abnormality of brain parenchyma. Axial T2-weighted spin echo MR image (B) at circle of Willis and coronal T2-weighted spin echo MR image (C) showed fainting but signal void of the supraclinoid and left A1 (large arrowhead), M1 segments persist (small arrowhead).

occlusion of the right A1 segment (figure 5B) were demonstrated during right ICA study. Prominent enlargement with elongated perforators of the M1 segment was noted. Transdural supply from middle meningeal artery (MMA) to right and left ACA (figure 5C, D) was demonstrated. The posterior cerebral artery (PCA) was also narrowed and irregular, but to a lesser extent than bilateral ICA (figure 5E). The cerebral angiography suggested moyamoya syndrome.

Discussion

Moyamoya is a nonspecific radiologic syndrome [1] which is usually characterized by bilat-

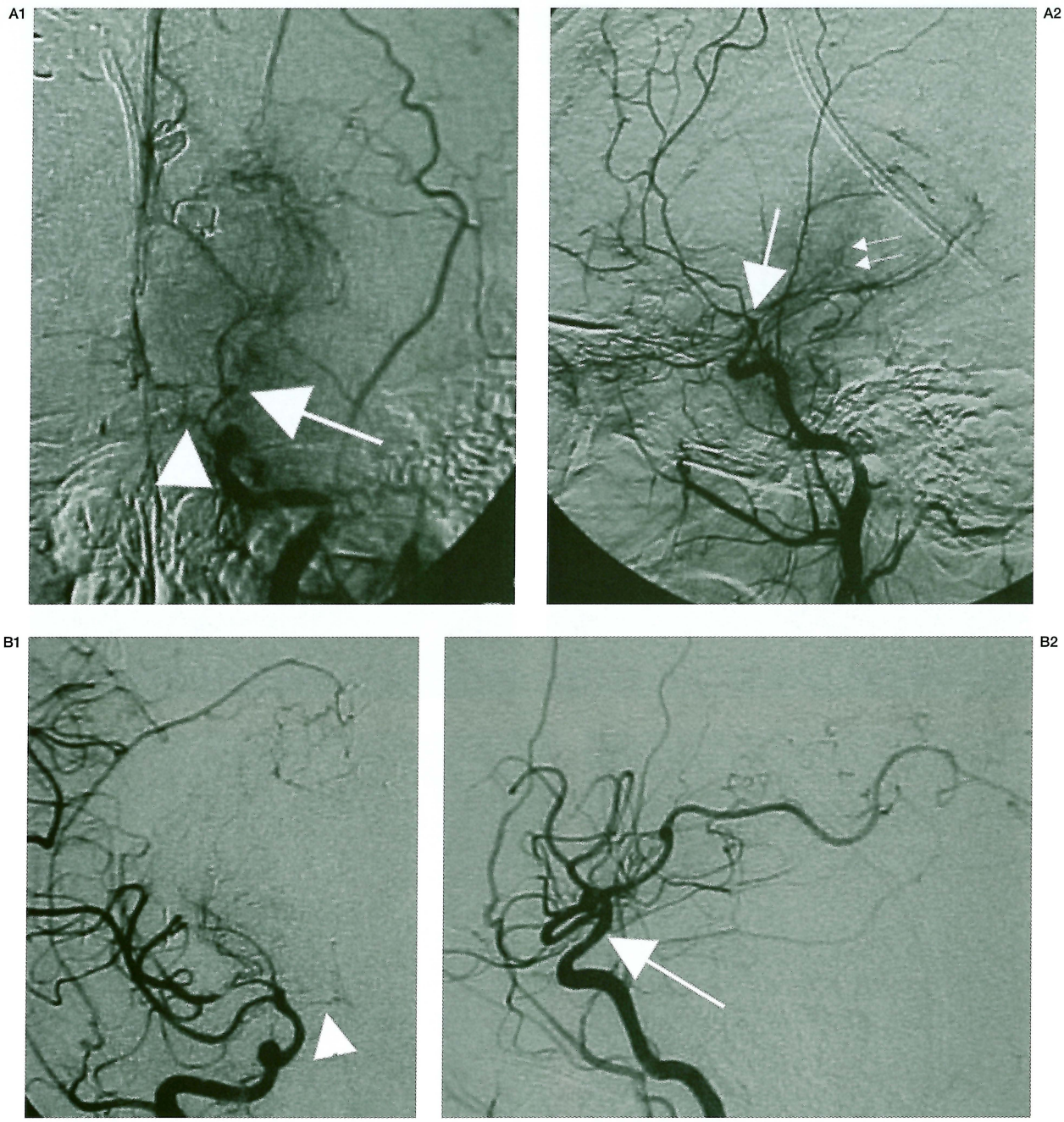

Figure 5

eral stenosis of C1 and/or C2 of the internal carotid artery and by a hazy network of the collateral circulation of the skull base.

This condition must be carefully observed in a patient who has had cranial irradiation. Many previous reports [1,7,8,10,11,12] stated that cranial irradiation in some circumstances, such as brain tumors which received radiation dose ranges as minimum as 4,000 rads, had developed large intracranial occlusive vasculopathy, moyamoya syndrome.

The clinical symptoms in children were headache, seizure and stroke [2] which can mimic tumor recurrence. Our patient, who had a germinoma of the pineal region, the most radiosensitive tumor with the best outcome after

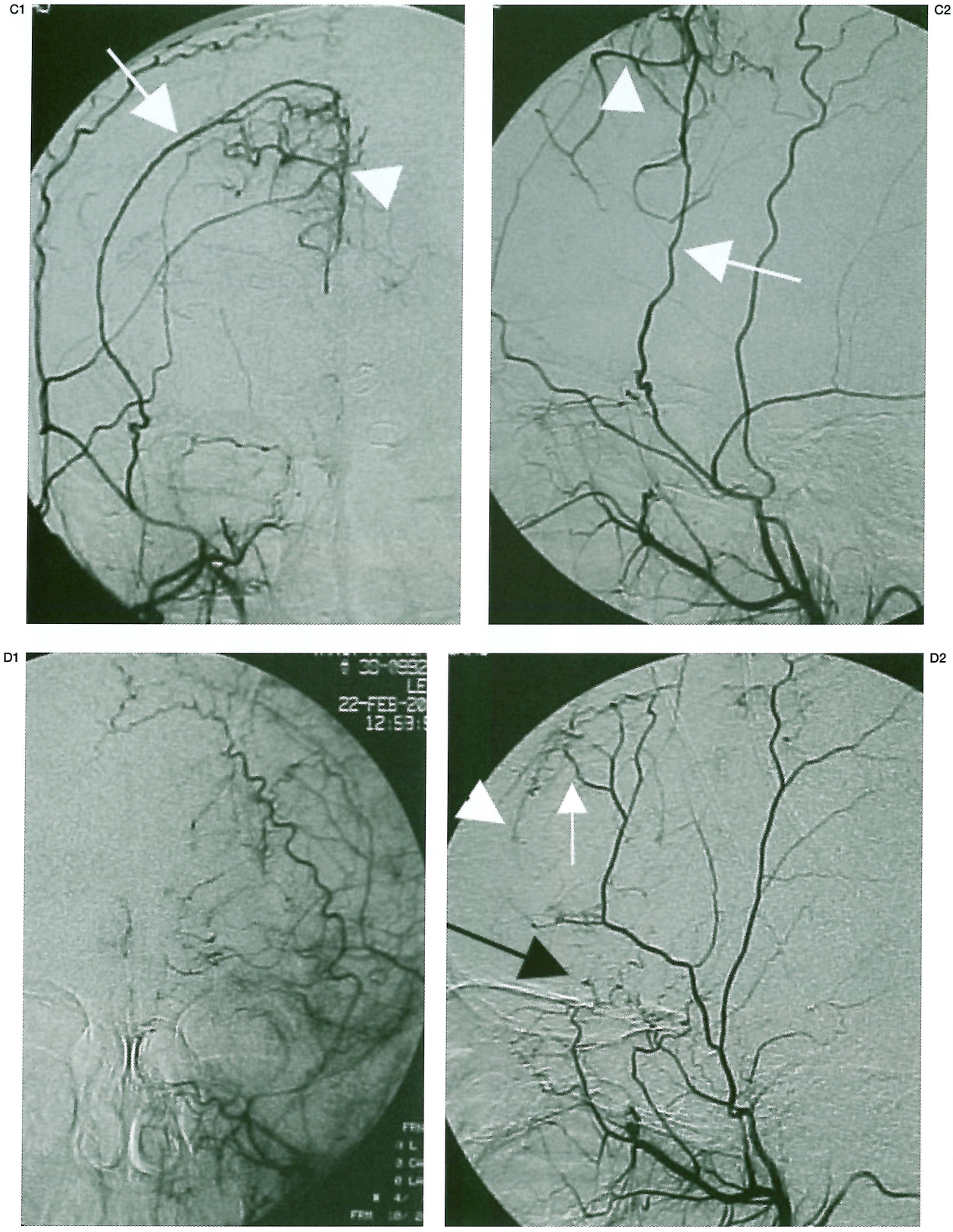

Figure 5

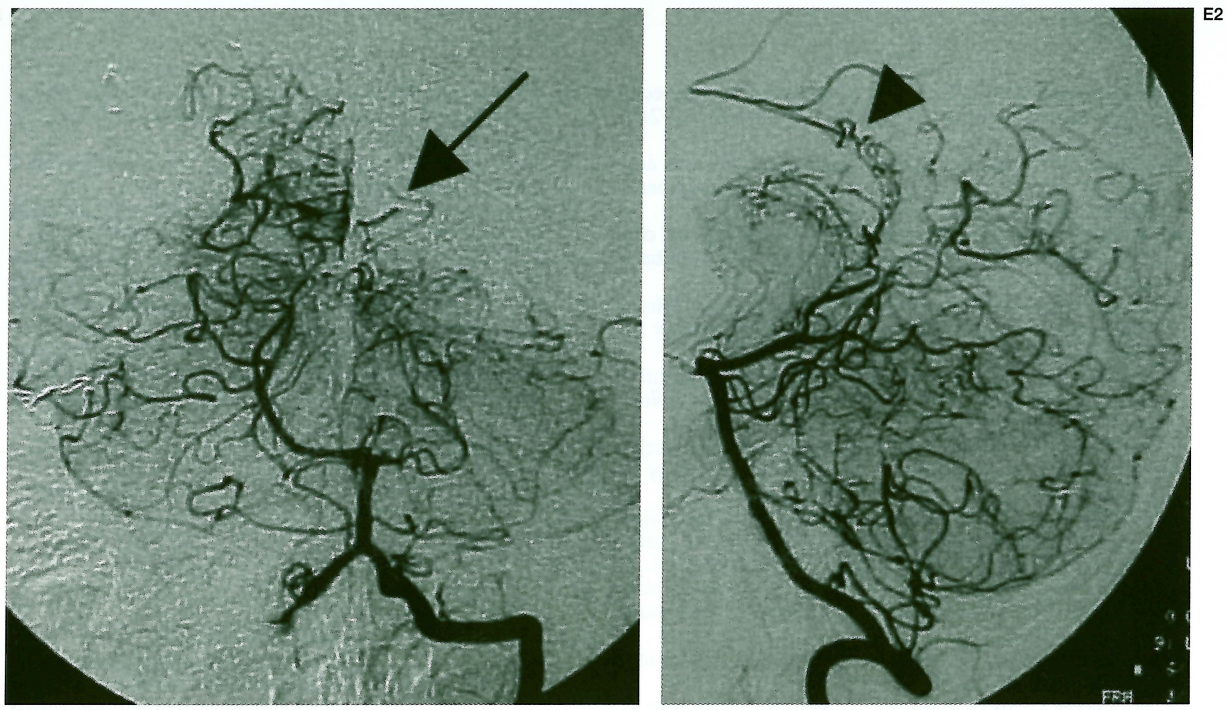

Figure 5 Cerebral angiography: Left ICA (A) frontal and lateral study revealed occlusion of the left supraclinoid part of ICA (arrow) and A1 (arrowhead), M1 segments with dilatation of the lenticulostriate arteries (double arrows). Right ICA (B) study demonstrated narrowing of the supraclinoid segment of the right ICA (arrow) and occlusion of the right A1segment (arrowhead). Right ECA (C) showed transdural collateralisation from right middle meningeal artery (MMA) (arrow) to right distal anterior cerebral artery (arrowhead). Left ECA (D) showed transdural anastomosis from left MMA (white arrow), superficial and deep temporal arteries to supply the brain (black arrow). Left VA study (E) Towne's and lateral projections showed occlusion of the left PCA (arrow), and pial-pial anastomosis (arrowhead) between posterior splenial artery and distal pericallosal artery.

radiation therapy [13]. He received a radiation dose of 5,620 rads, therefore having a chance of moyamoya syndrome development.

The patient developed new clinical symptoms five years after treatment when recurrent tumor could not be excluded, so that MR imaging was performed. MRI can detect tumor recurrence and the signal void of moyamoya vessels appearing as multiple, small round masses or tortuousity in the basal ganglia and thalamus [10].

The tumor was cured and MR imaging in this patient did not show evidence of moyamoya vessels in the basal ganglia but normal signal intensity of intracranial vessels at the circle of Willis, which can overlook the occurrence of moyamoya syndrome. Eight months later, when the patient developed stroke, cerebral angiography was performed. Angiography disclosed moyamoya syndrome composed of asymmetrical involvement of the distal ICA more on the left side, leptomeningeal anastomosis with transdural supply from the right deep temporal artery at the base of skull, right middle meningeal artery at convexity and posterior cerebral artery involvement.

Because moyamoya is a progressive disease and when developed clinical manifestations imply the collateral circulation is not sufficient, treatment intervention such as by-pass surgery [14] or encephalo-duro-arterio-synangiosis [15] may be needed. Even though MR imaging could detect moyamoya vessels [6] in early stages as in our patient, it was not sensitive enough.

We propose that cerebral angiography should be performed as soon as possible in patients who receive cranial irradiation and experience new symptoms which conventional imaging such as CT and MR cannot explain properly.

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